Paediatric Pathology Society

Proceedings of the Nineteenth Annual Meeting

The Nineteenth Annual Meeting was held in Glasgow on 19 and 20 October 1973, at the Royal Hospital for Sick Children.

Scientific communications

Hydatidiform mole as example of failed prenatal selection. D. I. Rushton. Department of Pathology, Birmingham Maternity Hospital, Edgbaston, Birmingham B15 2TG.

Placenta membranacea. J. Pryse-Davies. Bernhard Baron Research Laboratories, Queen Charlotte's Maternity Hospital, Goldhawk Road, London W6 0XG.

A case of placenta membranacea was described in a 16-year-old girl who bled in early pregnancy and required up to 10 l. blood by transfusion and aborted at 20 weeks' gestation. In retrospect, the condition could have been diagnosed by ultrasound examination at 11 weeks' menstrual age. (To be published elsewhere.)

Prognosis of germ cell tumours of the ovary in children. F. A. Langley and J. K. Steward. Department of Pathology, St. Mary's Hospital, Whitworth Park, Manchester M13 0JH.

Adrenal changes in stillborn infants. M. J. Becker. Laboratory of Pathological Anatomy, University of Amsterdam, Wilhelmina Gasthuis, Amsterdam, Holland.

From an extensive study of the causes of intrauterine death it evolved that the distribution of fat in the fetal zone of the adrenal cortex could give an indication of the mode of death of the fetus. Three different types of fat distribution were recognized.

Type I, characterized by absence of fat in the fetal zone, correlated with normal placentas and the clinical history of an acute complication. In contrast, type III, characterized by fatty transformation of the complete fetal zone, correlated with placentas with severe circulatory disturbances and a clinical picture of toxæmia, i.e. chronic fetal distress. Type II was an intermediate type, characterized by a fetal zone showing an irregular fatty transformation, with the main concentration around the central vein leaving a clear zone under the definitive cortex. Type II correlated with a subacute mode of death, like infections and blood group incompatibility.

The study showed that the pattern of fat distribution in the fetal zone of the adrenal cortex is indicative of the mode of death—acute, subacute, or chronic—and therefore may have clinical significance.

Myocardial infarction in the newborn. N. J. Brown. Department of Pathology, Southmead Hospital and Royal Hospital for Sick Children, Bristol BS10 5NB.

Myocardial infarction in infancy can be due to anomalous origin of the coronary artery. Much more rare is its occurrence when the origin of the coronary arteries is normal; 2 such cases were presented.

Twelve hours after normal birth the first infant became cyanosed and shocked with cardiac enlargement, baffling ECG, and evidence of nonfunctioning left ventricle. Myocardial infarction was suspected and he died next day. Necropsy revealed a massive recent anterior myocardial infarct. The coronary arteries were macroscopically normal, but histologically there appeared to be an embolus in the anterior descending branch. This could have been paradoxical embolism from the ductus venosus as suggested by Berry (1970).

The second infant developed heart failure when aged 5 hours with a highly abnormal ECG. Myocarditis was suspected and he died next day. Necropsy revealed a large recent anterior myocardial infarct. Histologically there was muscle necrosis but no myocarditis. The coronary arteries were normal apart from mild perivascular fibrosis of doubtful significance. Birth asphyxia, trauma, disseminated intravascular coagulation, arterial calcification, and supravalvular aortic stenosis were all excluded as aetiological factors and no satisfactory explanation of the infarct could be offered.

Reference


Transfer factor was administered to a 10-month-old infant with severe combined immunodeficiency disease in an attempt to stimulate cell-mediated immunity. No change in thymus-dependent lymphocyte function was observed. However, 3 weeks after transfer factor there was a marked increase in the leucocyte count with the appearance of mature plasma cells in the peripheral blood and bone marrow. Serum IgM rose to 2000 mg/100 ml; several blood group antibodies, including
anti-i, were now readily detectable. Though the relation of transfer factor to the polyclonal gammopathy may be coincidental, it is possible that in severe combined immunodeficiency disease transfer factor may induce uncontrolled B cell proliferation.

Biochemical state of the vitreous humour of infants at necropsy. P. G. F. Swift, E. Worthy, and J. L. Emery. Department of Pathology, Children’s Hospital, Western Bank, Sheffield S10 2TH. To be published in full in the Archives.

Hypernatraemia and uraemia in unexpected death in infancy. J. L. Emery, P. G. F. Swift, and E. Worthy. Department of Pathology, Children’s Hospital, Western Bank, Sheffield S10 2TH. To be published in full in the Archives.

Paediatric dosage of gentamicin. T. A. McAllister and D. G. Young. Department of Pathology, Royal Hospital for Sick Children, Yorkhill, Glasgow C.3.

Otoxicity of the broad spectrum antibiotic gentamicin has been recognized for 10 years and it has been customary to use it cautiously to achieve blood levels of 2 to 10 µg/ml. Most sensitive bacteria have a minimum inhibitory concentration of less than 2 µg/ml and otoxicity is most unlikely below 10 µg/ml. Evidence of gentamicin nephrotoxicity is poor unless it is used in combination with cephalosporins or diuretics.

Therapeutic levels are achieved in adults with a dose of 0·8 mg/kg 3 times daily, but with this regimen results in children are disappointing. The drug is excreted by glomerular filtration, which is proportional to surface area and therefore more efficient in children, especially neonates.

We studied 61 peak serum assays in 30 children with severe or potentially severe sepsis given 24 to 3 times the adult dose intramuscularly or intravenously. The children had a variety of surgical conditions, complicated by infection, and ranged in age from 3 days to 12 years. There was little difference in response in the different age groups. Apart from 3 'dampened responders', we concluded that children require a dose of at least 2 mg/kg 3 times daily for therapeutic efficacy. Neonates may require more, but all cases should be monitored by serum assays. These recommendations have been submitted to the Committee on the Safety of Medicines.

Adult type of polycystic disease of kidneys and liver presenting in childhood. B. G. Ockenden. Department of Pathology, North Staffordshire Royal Infirmary, Stoke on Trent.


Chromosome studies and the paediatric post-mortem. R. Sutherland, R. Bauld, and A. D. Bain. Department of Pathology, Royal Hospital for Sick Children, Sciennes Road, Edinburgh EH9 1LF.

Since October 1972 chromosome studies have been carried out on all stillbirths and infants coming to postmortem. The results of the study to date were presented and their significance discussed. The frequency of chromosome abnormalities in this group is between 5 and 10%. In view of the high incidence of chromosome abnormalities it is suggested that chromosome studies should be routinely carried out at the paediatric postmortem.

Rubinstein-Taybi syndrome. A. J. Barson. Department of Pathology, St. Mary’s Hospital, Whitworth Park, Manchester M13 0JH.

The case is described of a growth-retarded infant with multiple congenital anomalies born at 33 weeks’ gestation to a 30-year-old woman with 1 normal son, after a pregnancy complicated by hydranmios. The infant died on the 4th postnatal day with congestive heart failure, jaundice, and skin petechiae.

The infant had abnormally broad tips to the fingers, thumbs, and toes, an anomaly characteristic of the Rubinstein-Taybi syndrome. In addition he had a small maxilla and mandible with a high, arched, furrowed palate, a depressed nasal bridge with the septum below, low-set abnormal ears, palmar simian creases, an atrial septal defect, a 4-lobed right lung, biliary atresia, bilateral hydroureters, a large anterior fontanelle, deficient falx cerebri, and a Dandy-Walker type of malformation of the cerebellum. Histologically the thyroid and adrenal glands showed disordered differentiation.

These features are all compatible with the syndrome. As with previously reported cases, no chromosomal anomaly was shown.

Deformity of the lateral cerebellar lobes in children with meningomyelocele. S. Variend. Department of Pathology, Children’s Hospital, Western Bank, Sheffield S10 2TH.

A study was made of the gross structural deformity of the cerebellum from 100 children with meningomyelocele. The fissural pattern of the superior surface was classified in a variety of forms indicating degrees of displacement of the major fissures.

The portion of the cerebellum that had herniated into and through the foramen magnum was also described and classified. When the changes in the cerebellar tail were correlated with those of the upper and posterior surfaces, it appeared obvious that the deformity of the cerebellum is essentially a displacement backwards over the superior surface and downwards over the posterior surface.

It has already been shown that the weights of these deformed cerebella are less than that of normal infants, thus opposing theories of tissue overgrowth. When minor degrees of deformed organs occur there is a resemblance to the early fetal cerebellum, but the more
Proceedings: Polyclonal gammopathy and lymphoproliferation after transfer factor in severe combined immunodeficiency disease.

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Arch Dis Child 1974 49: 494-495
doi: 10.1136/adc.49.6.494-f

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